CASE REPORT

Mantle Cell Lymphona Masquerading as Obstructive Sleep Apnoea

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ABSTRACT

Tonsillar malignancy typically presents with asymmetrical tonsillar enlargement, lesion on the tonsils, sore throat or a neck mass. We report a case of unsuspected tonsillar malignancy in a 56-year-old gentleman who presented with symptoms of obstructive sleep apnoea. His tonsils were grade III bilaterally with normal mucosa. Tonsillectomy was performed to improve patient's compliance with Continuous Positive Airway Pressure (CPAP) therapy. These tonsillar specimens were reported to be Mantle Cell Lymphoma (MCL) based on the histology and ancillary studies. This case highlights that benign-looking symmetrical tonsillar enlargement can harbour occult malignancy. It is important to note that OSA symptoms may be the presentation for haematological malignancies. Tonsillar specimens should be sent for histopathological examination regardless of the indication to avoid misdiagnosis and delay in treatment.
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INTRODUCTION

Obstructive sleep apnoea (OSA) is a chronic disease characterised by episodes of complete or partial collapse of the airway associated with reduction in oxygen saturation. The commonest cause of OSA is obesity, however, head and neck tumour can be a potential aetiology. Diffuse large B-cell lymphoma is the commonest type of tonsillar lymphoma. There is limited literature found on Mantle Cell Lymphoma masquerading as OSA. The purpose of reporting this case is to raise awareness among clinicians that benign looking tonsils may also harbour occult malignancy. Therefore, histopathological examination of tonsils is essential to avoid missing an important diagnosis.

CASE REPORT

A 56-year-old gentleman presented to Otorhinolaryngology (ORL) clinic with snoring with choking episodes during sleep for the past three years. This resulted in severe excessive daytime somnolence with the Epworth Sleepiness Scale (ESS) score of 18. He did not have history of recurrent tonsillitis or any constitutional symptoms that may suggest underlying malignancy. Patient has a normal BMI of 23kg/m2 with no underlying co-morbidities. His tonsils were Grade III bilaterally with normal overlying mucosa. Polysomnography (PSG) confirmed the diagnosis of severe OSA with the Apnoea-Hypopnoea Index (AHI) of 50.6/hour, and minimal Spo2 of 76%. Subsequently, Continuous Positive Airway Pressure (CPAP) was given as the treatment of choice.

Patient had poor compliance to CPAP therapy due to the high-pressure requirement. Tonsillectomy was performed to improve the compliance. Histopathology examination (Figure 1) showed malignant lymphoid tumour, characterised by expanded mantle zones and focal diffuse growth pattern. The tumour composed of small to medium sized lymphoid cells with irregular nuclear contour. The malignant lymphoid cells were immunoreactive for CD20, CD79a and BCL2, cyclin D1 and CD5. Ki67 showed a low proliferative index (5%). The diagnosis of Mantle Cell Lymphoma (MCL) was made.

Postoperative recovery was uneventful. The CT scan (Figure 2) showed multiple cervical and abdominal lymphadenopathy of sub-centimeter in size. He was referred to Haemato-Oncology clinic for further management. His white cell count was 6 k/uL with Lactate Dehydrogenase (LDH) level of 599U/L. The disease burden was stratified as "low" according to

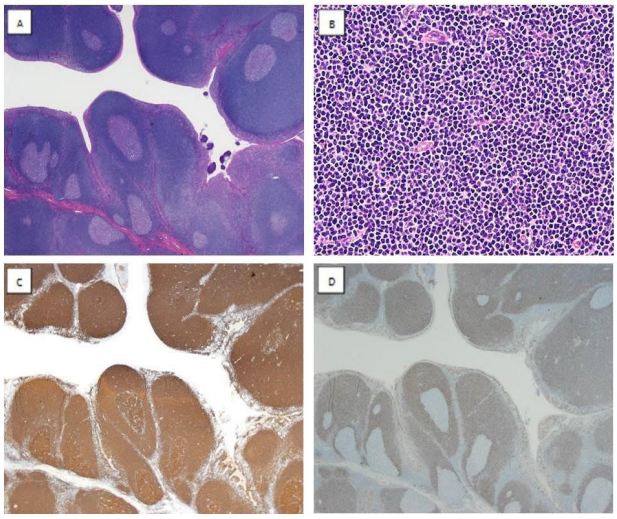

Figure 1: (A) Mantle cell lymphoma, expanded mantle zones and focal diffuse growth pattern. Residual germinal centres with tingible body macrophages are identified (Hematoxylin & Eosin, x20). (B) The tumour cells are monotonous, small to medium sized with irregular nuclear contours (Hematoxylin & Eosin, x400). (C) The tumour cells show diffuse and strong CD20 expression (Immunohistochemical stain, x20). (D) The Cyclin D1 immunostain shows nuclear positivity (Immunohistochemical stain, x20).

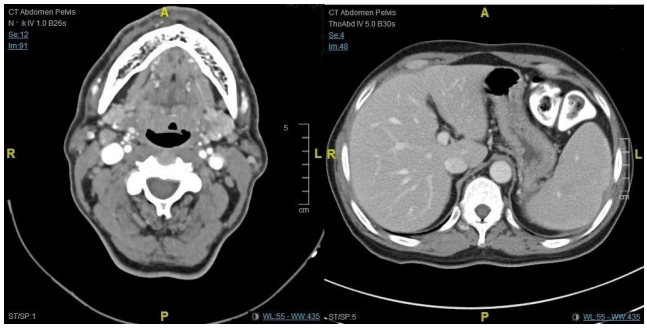

Figure 2: (A) Contrasted CT scan of the neck showed multiple enhancing and enlarged cervical lymph nodes at level Ib and level II. (B) Contrasted CT scan of the abdomen showed multiple subcentimeter lymph nodes at peripancreatic and para-aortic regions with splenomegaly.

Mantle Cell International Prognostic Index (MIPI) score. He was put on a "watchful waiting" treatment strategy and regular follow-up with blood investigations every three monthly. Chemotherapy is only indicated if he became symptomatic or presence of evidence of moderate to high MIPI score.

DISCUSSION

OSA is defined as AHI of five or higher associated with excessive sleepiness and arousal from sleep. Epworth Sleepiness Scale (ESS) is the best available tool to guide clinicians as to the patient's perception of sleepiness. Ear, nose, throat (ENT) and neck examinations are necessary to assess the patient's anatomical preponderance to OSA. Occasionally, these examinations may reveal some underlying pathological findings that have been masked as OSA. There have been reported cases of OSA as the first manifestation of chronic lymphocytic leukaemia (1).

In this case report, the patient presented with symptoms of OSA with high ESS score and diagnosis was confirmed with polysomnography. CPAP is the treatment of choice for patients with moderate to severe OSA. Nevertheless, non-compliance is a major issue in patients with CPAP therapy. High pressure requirement of the CPAP due to structural obstruction often cause discomfort, hence affects compliance. In this case, tonsillectomy was performed to reduce the pressure requirement of CPAP to improve the compliance.

To date, it is still controversial whether routine tonsils specimen require histopathological examination (HPE). Irfan, et al. reported that no specimen was found to be malignant in his recent audit of total 197 patients who had undergone routine tonsillectomy (2). Most specimens were reported as "reactive lymphoid hyperplasia "and "lymphoid follicular hyperplasia". There was no unsuspected tonsillar malignancy throughout the study period. Hence they advocated that routine HPE for tonsil specimen is not clinically justified unless malignancy is suspected. Several risk factors have been identified for tonsillar malignancy such as prior history of head and neck cancer, tonsillar asymmetry, visible lesion on the tonsils, neck mass or constitutional symptoms (3). In this case report, the patient had none of the risk factors mentioned. It is however a routine practice in our centre to send tonsillar specimens for HPE regardless of the indication. Much to our surprise, HPE and immune-histochemical studies of these tonsil specimens confirmed the diagnosis of Mantle Cell Lymphoma (MCL). The diagnosis would have been missed if the tonsils specimen were not sent for histopathology examination.

Despite an extensive literature search, there were limited cases of MCL reported. WHO has classified MCL as a rare distinct type of lymphoma with aggressive behaviour with an indolent course. The median age of presentation is 60 years old with male predominance. It represents about 4% of all lymphoma in the US and 7-9% in Europe. Patient with MCL normally presents with extensive cervical lymphadenopathy, splenomegaly, blood and bone involvement. Other possible extra-nodal sites involvement include stomach, colon, liver, lacrimal gland and Waldeyer's ring (4).

The diagnosis of MCL is made by histology examination coupled with ancillary studies of the biopsy material. Histologically, the tumour is composed of monomorphic small to medium sized lymphoid cells with an irregular contour. Almost all of the cases were immune-reactive for cyclin D1with CCND1 translocation detected. Further staging procedures include a complete blood

count, lactate dehydrogenase (LDH) level and CT scan of the thorax, abdomen and pelvis (5). Mantle cell International Prognostic Index (MIPI) is formulated to indicate the prognostic factors for overall survivor rate based on age, ECOG performance status, LDH level and white cell count. The treatment of MCL also depends on the MIPI score. The patient in this report has a low MIPI score, watchful waiting with regular monitoring of the disease progression was the treatment of choice.

CONCLUSION

This case report clearly highlights that OSA patient with bilateral enlargement of tonsils should be thoroughly evaluated for potential occult tonsillar malignancy. Snoring is no longer a normal behaviour, it is a sign of an underlying disease until proven otherwise.

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